PAPER

Parkinson's disease with camptocormia

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Background: Camptocormia is defined as an abnormal flexion of the trunk that appears when standing or walking and disappears in the supine position. The origin of the disorder is unknown, but it is usually attributed either to a primary or a secondary paravertebral muscle myopathy or a motor neurone disorder. Camptocormia is also observed in a minority of patients with parkinsonism.

Objective: To characterise the clinical and electrophysiological features of camptocormia and parkinsonian symptoms in patients with Parkinson's disease and camptocormia compared with patients with Parkinson's disease without camptocormia.

Methods: Patients with parkinsonism and camptocormia (excluding patients with multiple system atrophy) prospectively underwent a multidisciplinary clinical (neurological, neuropsychological, psychological, rheumatological) and neurophysiological (electromyogram, ocular movement recording) examination and were compared with age-matched patients with Parkinson's disease without camptocormia.

Results: The camptocormia developed after 8.5 (SD 5.3) years of parkinsonism, responded poorly to levodopa treatment (20%) and displayed features consistent with axial dystonia. Patients with camptocormia were characterised by prominent levodopa-unresponsive axial symptoms (ie, axial rigidity, gait disorder and postural instability), along with a tendency for greater error in the antisaccade paradigm.

Conclusion: We suggest that (1) the salient features of parkinsonism observed in patients with camptocormia are likely to represent a specific form of Parkinson's disease and camptocormia is an axial dystonia and (2) both camptocormia and parkinsonism in these patients might result from additional, non-dopaminergic neuronal dysfunction in the basal ganglia.

Camptocormia is defined as an abnormal flexion of the trunk that appears when standing or walking and disappears in the supine position (fig 1). This rare symptom, usually found in patients >65 years of age, is often attributed to normal ageing and is not diagnosed. The term ''camptocormia'' is derived from the Greek words ''kamptos'' (to bend) and ''kormos'' (trunk), and was coined by the French neurologist Souques.[1] The authors reported a soldier, who had a forced posture with a bent-forward trunk after a gunshot wound. The patient was reportedly cured after a psychotherapeutic interview and subsequent application, in narcosis, of a plaster jacket that was removed after 2 days. Souques even suggested that the disorder could be cured completely and permanently by means of a persuasive galvanisation or faradisation of the dorsolumbar region, and termed the condition ''cyphose hystérique'' (''hysterical kyphosis''). Although a few authors pointed out that patients with an organic condition such as spondylitis should be differentiated from those with hysterical kyphosis, a psychogenic explanation for the disorder prevailed, owing to the many cases similar to those of Souques that were reported during the First and Second World Wars.[2–6]

In 1995, Laroche *et al*[7] reported 37 patients with camptocormia who were compared with 15 age-matched patients without camptocormia but with posterior interapophysial osteoarthritis and elderly patients surgically treated for narrowing of the lumbar canal. In patients with camptocormia, magnetic resonance imaging (MRI) showed features consistent with circumscribed myopathy in the paravertebral muscles, and it was stated that camptocormia might be ascribed to a primary paravertebral myopathy.[8] The spectrum of these neuromuscular disorders was subsequently extended to myasthenia gravis, nemaline myopathy,[9 10] amyotrophic lateral sclerosis,[11] inclusion body myositis, polymyositis[9 12] and miscellaneous causes such as paraneoplastic disorder[13] and valproate toxicity[14] (see also review by Azher and Jankovic[15]). None of the above studies, however, mentioned the presence of parkinsonism. Indeed, in the few studies that have dealt with the issue of camptocormia and parkinsonism, the coexistence of these disorders was thought to result from the incidental occurrence of Parkinson's disease and neuromuscular disorders,[9 15 16] features of multiple systemic atrophy (MSA),[9 15] or a rare type of dystonia peripherally induced[15] or of unknown origin.[9 15 17] Although some studies were the first to draw attention to the disorder,[17] the nosological position and the origin of this unusual symptom in association with parkinsonism remain unclear, as do the precise clinical features of this entity.

The aim of the present prospective study was to characterise a selected sample of patients with parkinsonism and camptocormia and to compare them with age-matched patients with Parkinson's disease without camptocormia.

PATIENTS AND METHODS

Patients

Sixty three patients with Parkinson's disease examined in our institution between January 1995 and December 2002 met the criteria for inclusion in our study—namely: (1) age >30 years; (2) presence of camptocormia, defined as an anterior flexion of the thoracolumbar spine from 15° to 90° appearing in orthostatism or after gait and disappearing in the recumbent position; and (3) parkinsonism, defined as the presence of an akineto-rigid syndrome with or without

Abbreviations: MRI, magnetic resonance imaging; MSA, multiple systemic atrophy; UPDRS, Unified Parkinson's Disease Rating Scale

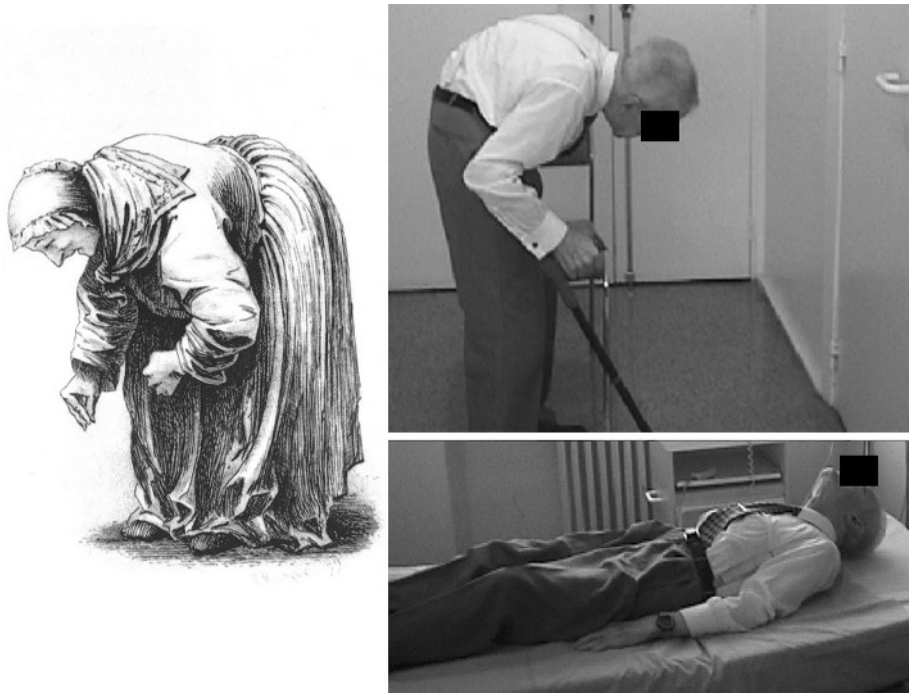

Figure 1 Drawing of a patient with Parkinson's disease and camptocormia (Bibliothèque Charcot, Hôpital de la Salpêtrière; left) and photographs of one of the patients studied (camptocormia appears when the patient is standing or walking and disappears in the supine position; right). Patient consent was obtained for publication of this figure.

tremor.[18] Eleven of these patients could not be contacted (n = 7) or were deceased (n = 4), and a further 17 refused to participate. Of the remaining 35 patients, 18 were not included in the study because of the presence of an exclusion criterion: (1) presence of an identified cause of camptocormia such as myopathy (observed in one patient and confirmed by biopsy), motor neurone disease, primary or secondary dystonia, or spinal cord disease on MRI; (2) history of spinal surgery on more than one occasion (n = 1) and presence of osteoarticular lesions on spinal cord MRI: arthrodesis (n = 3), more than three different herniated discs (n = 2) or scoliosis with Cobb's angle >30° (n = 3); (3) presence of neurological symptoms suggestive of probable MSA (n = 2)[19]; and (4) dementia (mini-mental status[20] <24, n = 6). In all, 17 patients (6 women, 11 men) completed the study, and none had evidence of motor neurone disease, primary or secondary generalised dystonia or camptocormia of psychogenic origin. Age at inclusion, age at onset and duration of parkinsonism were (mean (standard deviation (SD)) 69.3 (7.2), 57.0 (10.5) and 12.2 (6.9) years, respectively. The mean (SD) Hoehn and Yahr ''off'' score[21] was 3.3 (0.9) and mean levodopa-equivalent dosage[22] was 985 (535.4) mg/day.

To compare the distinguishing traits of patients with Parkinson's disease with and without camptocormia, 8 of the 17 patients with camptocormia were matched for age, disease duration and severity of the parkinsonian syndrome (Hoehn and Yahr ''off'' drug score) with 8 levodopa-responsive patients with Parkinson's disease without camptocormia (recruited in parallel, who accepted to participate in the study; table 1). All the patients gave written informed consent in accordance with the Declaration of Helsinki, and the protocol was approved by the local ethics committee of the Groupe Hospitalier Pitié-Salpêtrière (Paris, France).

Clinical evaluations

Patients with and without camptocormia were evaluated in the same fashion. The severity of camptocormia was scored in the ''off'' and ''on'' drug conditions as follows: the patient was instructed to stand with his or her back to a wall without attempting to stand upright, and the distance between the seventh cervical vertebra and the wall was measured. Camptocormia-related disability was measured using the Echelle d'Incapacité Fonctionnelle pour l'Evaluation des Lombalgies (EIFEL) questionnaire,[23] a validated French version of the Roland–Morris Disability Scale.[24] The Roland–Morris Scale is a valid and reliable 24-item measure of pain-related disability derived from the Sickness Impact Profile.[25] The various items concern functional limitations for various activities, seeking help from others and changes in affect, appetite and sleep due to pain. Patients were requested to select any item that currently applied to them. The scale is scored from 0 to 24, with a higher score indicating more severe disability. Although developed as a measure of physical disability for low back pain, the Roland–Morris Scale, reworded without reference to the back, has been found to be a reliable and valid measure of physical disability for patients with other chronic pain problems.[26] Finally, a physical examination was carried out, with attention focused on camptocormia-related osteoarticular and muscular changes.

Table 1 Characteristics of matched patients with Parkinson's disease with and without camptocormia

	With camptocormia (n = 8)	Without camptocormia (n = 8)
Age (years)	68.2 (8.7)	67.1 (8.0)
Age at onset of parkinsonism (years)	57.9 (8.4)	56.7 (8.4)
Disease duration (years)	10.4 (4.7)	10.7 (4.8)
Hoehn and Yahr score (off)	3.4 (0.9)	3.3 (0.9)
Levodopa equivalent (mg/day)	794 (453)	794 (253)

Values are mean (SD).

The percentage improvement in activities of daily living (ADL-Unified Parkinson's Disease Rating Scale (UPDRS) part II[27]) was determined with respect to the ''off'' drug condition. Evaluations of the modified motor disability score (UPDRS part III without item 28 = posture[27]), the ''modified'' axial score (defined as the sum of the following motor subscores: speech, facial expression, neck rigidity, gait and postural stability; items 18, 19, 22a, 29 and 30 of UPDRS part III) and the Martinez-Martin gait score[28] were performed in the ''off'' state—that is, after an interruption of at least 12 h after giving drugs for Parkinson's disease (Core Assessment Program for Surgical Interventional Therapies[29]), and in the best ''on'' drug condition after the administration of a single suprathreshold dose of levodopa (50 mg higher than the usual effective dose taken in the morning). Levodopa-related complications were evaluated using UPDRS part IV.[27]

The neuropsychological assessment, carried out in the ''on'' drug condition, included (1) the Mini-Mental State Examination[20] and the Mattis Dementia Rating Scale[30] to evaluate global intellectual efficiency; (2) the Grober and Buschke test[31] to evaluate verbal learning; (3) the ''frontal'' score,[32] including the simplified version of the Wisconsin Card Sorting test[33] to evaluate rule generation, shifting abilities and attentional control, verbal fluency tests, the graphic series of Luria and evaluation of behavioural abnormalities such as inertia, indifference, prehension, imitation and use; (4) the Stroop Test[34] to estimate the inhibition of interference; (5) the Trail Making Test[35] to evaluate set-shifting; and (6) aspects of motor control, assessed by ''conflicting instructions'', evaluating resistance to interference, the ''go–no go test'' evaluating inhibitory control, ''Luria motor sequences'' and rhythm reproduction[36] evaluating motor programming and executive control of action.

Neuropsychiatric features were assessed using both a semistructured psychiatric interview (Mini-International Neuropsychiatric Interview 5.0.0[37]) and the Hospital Anxiety and Depression Scale.[38]

Laboratory tests

Different parameters were studied to screen for localised or systemic diseases that could be a possible cause of bent spine, including muscular (creatine protein kinase, lactate dehydrogenase, lactate, pyruvate, aldolase and carnitine plasma concentrations) and osteoarticular (albuminaemia, calcium and phosphorus, vitamin D) diseases. Hormonal levels (cortisol, thyroid and parathyroid hormones) and inflammatory (erythrocyte sedimentation rate) indices were also investigated. These blood tests were normal, except in one patient with camptocormia and pre-existing hypothyroidism whose raised thyroid-stimulating hormone level resolved after adjustment of the thyroxine dose.

The electrophysiological study was carried out with a Nicolet Viking IV (Nicolet Biomedical, Madison, Winconsin, USA). Sensory nerve conduction studies were carried out in the sural nerve. Motor nerve conductions studies were carried out in the tibial nerve and common peroneal nerve, with F wave analysis in both lower limbs. Distal motor latency, compound muscle action potential amplitude and motor nerve velocity were recorded. Neuromuscular transmission was tested by applying repetitive nerve stimulation (10 stimuli at 3 Hz) to the ulnar and accessory nerve. Myopathic and neuropathic changes were searched for using needle recording from several muscles (tibialis anterior, deltoid, rectus abdominis, psoas major, cervical and thoracolumbar paraspinal muscles).

Eye movements were recorded using horizontal binocular direct current electro-oculography as previously described.[39] The latency and velocity of horizontal visually guided saccades were studied with a gap paradigm. Subjects were instructed to fixate the central point and then look at the lateral target as soon as it appeared 25° randomly right or left. The mean saccade latency was calculated by averaging the values of 18 saccades in each direction. The ability to inhibit visually guided saccades was studied with the antisaccade paradigm[40]; the percentage of errors (misdirected saccades—ie, saccades either reaching or initially directed towards the target) was determined by 18 trials in each direction. We controlled vertical eye movements to rule out supranuclear palsy. All paradigms were conducted during the same session.

Statistical analysis

Demographic, clinical and laboratory parameters were described for the 17 patients with camptocormia as mean (SD). Patients with camptocormia were divided into different subgroups according to the medial value of the severity, duration and levodopa sensitivity of the camptocormia. Differences between these subgroups were analysed using the Wilcoxon test. For correlations between quantitative variables, the Spearman correlation test was used. The eight patients with Parkinson's disease with camptocormia and the eight matched patients with Parkinson's disease without camptocormia were compared using paired Wilcoxon rank sum tests. p Values <0.05 were considered significant. All tests were two sided. Statistical analyses were carried out using SAS V.8.1.

RESULTS

Characteristics of camptocormia

The 17 patients with Parkinson's disease developed camptocormia at a mean (SD) age of 65.5 (7.3) years and, at the time of the study, patients had had a bent spine for 3.8 (3.1) years. The onset of camptocormia was fairly rapid and progressive (a couple of weeks) in 70% of patients, and 80% of the patients experienced dorsolumbar back pain. None of the patients had a history of low back pain before the onset of camptocormia. The disorder never preceded the parkinsonism, but emerged concomitantly in one patient; in the other patients, it began a mean (SD) of 8.5 (5.3) years after the onset of parkinsonism. The severity of the bent spine as measured by the distance C7–wall was 19.5 (9.4) cm when the patients were examined without treatment for Parkinson's disease (''off'' drug) and decreased by 20% with the levodopa intake (''on'' drug distance C7–wall = 15.0 (5.6) cm). The mean level of disability resulting from camptocormia, as assessed using the EIFEL scale (from 0 (minimal disability) to 20 (severe disability)), was 9.7 (6.2). Camptocormia consisted of an anterior flexion of the thoracolumbar spine of at least 45° for each patient; it was accompanied by laterodeviation in five patients, two of whom presented with severe rubbing ulcers in the concavity of the deviation as a result of the very narrow angle made by the superior and external portion of the iliac crest and the chest. There was no antecollis or head drop, but bent spine was accompanied by a moderate (n = 8) to severe (n = 2) weakness of the gluteus maximus. In all, 35% of the patients with camptocormia had a hip flexion, and 45% a genu flexion, both of which were absent in the patients without camptocormia.

When patients were subdivided into those with severe and those with moderate camptocormia, the first had worse motor disability (modified ''on'' drug UPDRS III 33.2 (7.5) *v* 21 (8); $p<0.03$), but no difference was observed among the patients with camptocormia when subdivided with respect to the degree of levodopa responsiveness or the duration of camptocormia (not shown). The delay in the appearance of camptocormia was negatively correlated with the age of the

patient at onset of the parkinsonian syndrome. In brief, the older the patient at the onset of parkinsonism, the shorter the subsequent delay before the onset of camptocormia ($r = -0.63$; $p<0.005$).

Patients walked slowly (n = 14), with short steps (n = 13), and without swinging arms (n = 17), but tended to drag their legs in only three cases. Frequent falls and freezing were observed in 11 and 13 patients (start hesitations, n = 5; falls due to freezing, n = 5).

Comparison of clinical characteristics of the eight patients with Parkinson's disease with camptocormia and the eight patients with Parkinson's disease without camptocormia

Parkinsonian motor disability without treatment for Parkinson's disease was not different between patients with and patients without camptocormia. In the ''on'' drug condition, parkinsonian motor disability was more severe in patients with camptocormia compared with patients without camptocormia (table 2).

The severity of axial symptoms was not different between the two groups of patients in the ''off'' drug condition. The residual modified axial score (''on'' drug) was more severe in patients with camptocormia than in those without camptocormia, owing to a higher score for dysarthria, neck rigidity, postural stability and gait (items 18, 22a, 29 and 30 of the UPDRS part III; table 2). The Martinez–Martin gait disability score was improved in the ''on'' condition but was higher in patients with camptocormia in both the ''off'' and ''on'' drug conditions (''off'' drug condition: not significant; ''on'' drug condition: $p<0.02$; table 2).

The severity of the levodopa-related complications (UPDRS IV) was not different in patients with Parkinson's disease with and without camptocormia (table 2). The patients with camptocormia tended to have less levodopa-induced dyskinesias (item 32, UPDRS IV, 30% *v* 75%; not significant) and fewer daily motor fluctuations than patients without camptocormia (item 39, UPDRS IV, 25% *v* 62.5%; not significant; not shown).

We found no significant differences between patients with and without camptocormia with respect to the neuropsychological and neuropsychiatric evaluations (table 3).

Electrophysiological recordings

Nerve conduction studies showed only non-specific neurogenic changes in three patients with camptocormia. No myopathic abnormalities were detected in the deltoid, rectus abdominis, iliopsoas, cervical or thoracolumbar paraspinal muscles, nor were there any features consistent with a dysfunction of the neuromuscular junction.

For electro-oculographic recordings, as there were no differences between rightward and leftward values, all parameters were pooled electro-oculographic recordings could not be taken (in two patients with camptocormia and in one without camptocormia; table 4).

The average value of horizontal gaze velocity was within the normal limits in both groups of patients and not significantly different between patients with and those without camptocormia. More than 28% of the patients with camptocormia presented abnormal horizontal gaze velocities (ie, <230°/ms) compared with none of the patients with Parkinson's disease without camptocormia. Similarly, 43% of the patients with camptocormia made frequent errors in the ''antisaccade'' paradigm, whereas none of the patients with Parkinson's disease without camptocormia had abnormal results (not significant).

Table 2 Motor disability in matched patients with Parkinson's disease with and without camptocormia

	With camptocormia (n = 8)	Without camptocormia (n = 8)
Motor disability*		
Off	34.5 (13.6)	34.7 (15.5)
On	24.7 (11.5)†‡ [32]	13.3 (6.1) [58]
Axial motor score		
Off	9.9 (3.4)	7.5 (3.9)
On	8.1 (2.6)†‡ [22]	3.7 (1.5) [50]
Gait disorders[31]		
Off	40.1 (13.3)†	24.6 (16.4)
On	36.4 (8.8)†‡ [11]	10.2 (7.4) [56]
Motor complications[30]	3.0 (4.2)	4.6 (2.7)

Values are mean (SD). Percentage improvement compared with the ''off'' drug condition are in square brackets.
*Modified Unified Parkinson's Disease Rating Scale III (see Methods).
†$p<0.05$; comparison between ''on'' and ''off'' values in patients with camptocormia.
‡$p<0.05$; comparison between patients with and without camptocormia.

Table 3 Neuropsychological and psychiatric characteristics of matched patients with Parkinson's disease with and without camptocormia

	With camptocormia (n = 8)	Without camptocormia (n = 8)
Education (years)	11.4 (4.9)	8.6 (3.4)
Neuropsychological status		
Mattis' dementia rating scale[33]	133.6 (9.4)	134.7 (7.4)
Grober and Buschke memory test[34]	43.3 (8.1)	44.5 (3.3)
Frontal battery[35]	40.6 (8.8)	46.9 (8.1)
Psychiatric status		
Depression[41]	7.4 (2.5)	8.4 (5.0)
Anxiety[41]	8.0 (3.5)	10.9 (3.4)

Values are mean (SD).

DISCUSSION

To the best of our knowledge, this is the first case–control study on a selected sample of patients with parkinsonism and camptocormia compared with matched patients with Parkinson's disease without camptocormia. This enables us to define the clinical characteristics of both camptocormia and parkinsonian symptoms in this rare disorder compared with the classic form of Parkinson's disease.

The patients with camptocormia included in this study were rigorously selected, and differed from patients in other studies in which the occurrence of camptocormia with parkinsonian syndrome prompted a diagnosis of ''parkinsonian plus'' syndromes such as MSA. None had corticospinal

Table 4 Oculomotor characteristics of matched patients with Parkinson's disease with and without camptocormia

	Patients with camptocormia (n = 7)	Patients without camptocormia (n = 7)
Velocity of horizontal saccades		
Mean (SD) value (ms)	279.6 (66.1)	354.2 (64.7)
No of patients with abnormal results (<230°/ms)	28.5%	0%
Antisaccade paradigm		
Percentage of errors	30 (4–69)*	16.8 (4–25)*
Number of patients with abnormal results (>25%)	42.9	0

Values in parentheses are the limits of normal values calculated as 2 SD below or above the average control values of each test.
*Values in parentheses are minima–maxima.

signs, cerebellar signs or severe autonomic complaints, features that would have pointed towards a diagnosis of MSA. Furthermore, antecollis, another feature that should be distinguished from camptocormia, but which is reported to co-occur with camptocormia and could point to a diagnosis of either MSA[9 19] or cervical myopathy,[9] was absent in our patients. Finally, there was no evidence to suggest a diagnosis of inflammatory or endocrinometabolic myopathic causes of the camptocormia,[9 12 16] and the results of both the laboratory and electrophysiological nerve and muscle studies failed to detect any features suggestive of myopathic changes.

Most patients reported that camptocormia was more severe with action—namely while standing or walking—and tended to increase with time and fatigue during the day. This feature, along with the fact that this postural disorder of the trunk disappeared in the supine position, is consistent with the definition of axial[41] or action dystonia.[42] None of the patients had, however, a history of multiple lumbar disc hernia or laminectomy[43] (which would have suggested a peripherally induced axial dystonia[15]), although camptocormia developed in a rapidly progressive manner with mild to severe back pain in several patients. These results are consistent with those of Djaldetti *et al*,[17] who reported that camptocormia was painless in five of eight patients with Parkinson's disease with camptocormia.

Despite the fact that there was a negative correlation between the delay in the appearance of camptocormia and the age at onset of the parkinsonian symptoms, which may suggest that ageing is a contributive factor in the occurrence of camptocormia, the symptom never preceded the parkinsonian motor disability. This raises the question of whether the association of this axial dystonia with parkinsonism represents a distinct form of Parkinson's disease. However, parkinsonian motor disability was not different in patients with and without camptocormia (table 2). The response of patients with camptocormia to levodopa treatment was modest; they presented with severe axial symptoms and gait disorders, all symptoms known to respond poorly to levodopa.[44] Indeed, patients with dementia (n = 6) were excluded from the current study to remove dementia-associated brain lesions as a possible confounding factor correlating with axial features.[44] Taken together, the above-mentioned arguments suggest that the parkinsonian syndrome that accompanied the camptocormia is caused by selective degeneration of the nigrostriatal dopaminergic system together with distinctive additional non-dopaminergic brain lesions. Interestingly enough, camptocormia and parkinsonism have been recently reported as a phenotypic heterogeneity of a *parkin* mutation.[45] The fact that akinesia, rigidity and tremor present in patients with camptocormia responded—at least in part—to levodopa helps to distinguish these patients from those with atypical forms of Parkinson's disease, such as progressive supranuclear palsy, in which the cardinal parkinsonian symptoms are levodopa unresponsive.[46]

In line with this result, we found a tendency for a greater percentage of errors on the antisaccade paradigm in patients with camptocormia, as also reported in patients with progressive supranuclear palsy.[39] Interestingly, error commission in antisaccade paradigm is thought to result from dysfunction of one or several lesions within a network including the omnipause neurones in the brain stem, the rostral superior colliculus, part of the caudate and substantia nigra reticulata, and neurones in the frontal eye field and anterior cingulate cortex.[47] However, in contrast with patients with progressive supranuclear palsy,[39] around one third of patients with camptocormia seemed to present a change in horizontal ocular movement velocity. Although we cannot exclude that these results could reflect the extent of non-dopaminergic lesions in the basal ganglia, we suggest that our findings also point to neuronal dysfunction of the brain stem in these patients. Although the precise anatomical region involved is unknown, several anatomical, neurophysiological and pharmacological experiments suggest that the pedunculopontine might be implicated.[48 49]

In conclusion, the salient features of parkinsonism with camptocormia (axial rigidity, gait disorder, postural instability, poor levodopa responsiveness of axial symptoms) are likely to represent a selective form of Parkinson's disease in which a significant additional non-dopaminergic neuronal dysfunction occurs, both within the basal ganglia and the brainstem.

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